Coronary Arteriovenous Fistula Presenting as Chronic Pericardial Effusion

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In August 1998, the patient, a 75-year-old woman, was diagnosed with pericardial effusion (PE) during an investigation of cardiomegaly. The PE disappeared after the administration of diuretics, but in February 1999, shortness of breath and general fatigue developed, and PE was again present. Diagnostic pericardiocentesis revealed bloody fluid. Chest computed tomography revealed a markedly expanded and tortuous right coronary artery (RCA). Coronary angiography (CAG) confirmed a RCA–coronary sinus fistula, and there was a significant step-up of O2 saturation at the right atrium. Cardiac tamponade developed soon after CAG, so the patient underwent surgical closure of the CAVF. Although a bleeding point was not identified, the PE was disappeared after operation. Histopathologically, parts of the wall of the fistula were quite thin and erythrocytes and lymphocytes had infiltrated the pericardial space. The clinical course and the findings indicate that the CAVF caused chronic PE.

Key Words: Cardiac tamponade; Coronary aneurysm; Coronary arteriovenous fistula

Coronary arteriovenous fistula (CAVF) is an abnormal communication between a coronary artery and a cardiac chamber, great vessel, or other vascular structure. Most of them are found incidentally during coronary angiography (CAG), and are identified as a cause of a continuous murmur, myocardial ischemia, congestive heart failure or, rarely, bacterial endocarditis. However, cases with pericardial effusion (PE) caused by rupture of the aneurysmal coronary artery are quite rare; only 3 cases of PE caused by rupture of the CAVF have been previously reported. We report a case of chronic PE caused by a CAVF. The PE was observed for 6 months, and finally developed into cardiac tamponade.

Case Report

A 75-year-old woman was referred to hospital for investigation of recurrent PE. She had a history of pulmonary tuberculosis 50 years ago. In August 1998, she had been admitted to another hospital for further investigation of cardiomegaly (cardiothoracic ratio (CTR) 63%) and was diagnosed as having PE for the first time. However, she was free of symptoms and the laboratory examinations were normal, including thyroid function and tuberculin reaction. Chest computed tomography (CT) did not show any abnormal findings except for the PE and a scar from the pulmonary tuberculosis. She was treated with diuretics and the PE disappeared within 6 months. The CTR decreased from 63% to 54% on the chest X-ray. In February 1999, she developed shortness of breath and general fatigue, and echocardiographic examination revealed that the PE had recurred.

On the admission to hospital in February 1999, her vital signs were: blood pressure 148/70 mmHg, heart rate 92 beats/min, respiratory rate 20 breaths/min, and body temperature 36.5°C. Pulsus paradoxus was not detected. Chest auscultation revealed bilaterally normal respiratory sounds and a soft continuous murmur at the left sternal border of the 4th intercostal space. She had hepatomegaly, bilateral leg edema and a markedly expanded external jugular vein.

Laboratory tests were as follows: leukocytes 7.2 × 10^3/μl; C-reactive protein 0.5 mg/dl; erythrocyte sedimentation rate of 24 mm/h; TSH 1.86 IU/ml; free T3 1.77 pg/ml; free T4 0.83 ng/ml; tuberculin reaction 10 × 10 mm. A chest X-ray revealed cardiomegaly (CTR = 67%) without pulmonary infiltration. The electrocardiogram (ECG) on admission was normal except for flat T waves in leads V4–6 (Fig 1A). An enhanced CT scan revealed a dilated right coronary artery (RCA) and a dilated coronary sinus (CS) vein, an expanded inferior vena cava and a large, circumferential PE (Fig 2). However, leakage of contrast media into the pericardial space was not seen. A diagnostic pericardiocentesis yielded 20 ml of bloody fluid (Hct = 22%) that contained 5.7 mg/dl of protein, 56 mg/dl of glucose and 23.2 IU/L of adenosine deaminase. The culture for bacteria in the pericardial fluid was negative, and a polymerase chain reaction for the tuberculosis bacillus was not detected. There was no evidence of malignant cells. Coronary angiography confirmed a large (approximately 10 mm in diameter) and tortuous RCA, and contrast medium passed from the RCA to the expanded CS via a fistula (Fig 3). The left coronary artery was almost normal. Cardiac catheterization revealed a significant O2 step-up in the right atrium (RA) (O2 content: 70.4% in the superior vena cava, 85.4% in the RA). A left-to-right shunt and a Qp/Qs ratio were calculated as 62%...
and 2.5, respectively, and based on all these findings, we diagnosed a RCA–CS fistula.

One week after admission, the PE had progressively increased. The patient’s hemodynamic status rapidly deteriorated: systemic blood pressure decreased to 92/60 mmHg, RA pressure increased to 18 mmHg (mean), and heart rate increased to 100–130 beats/min in atrial fibrillation. Because of the development of cardiac tamponade, surgical closure of the RCA–CS fistula was immediately performed under cardiopulmonary bypass. Surgical exploration confirmed a large pericardial hematoma and approximately 300–400 ml of bloody fluid. The bleeding site, however, was not identified. The thrill of the shunt flow was palpable around the CS, and an aneurysmal dilatation was observed just distal of the posterior descending branch of the RCA. We therefore ligated both ends of the fistula. The resected aneurysmal wall of the fistula consisted of both arterial and venous structures. In the wall of the arterial structure, the elastic
fiber density was markedly decreased in the media and, furthermore, the internal elastic lamina was fragmented (Fig 4). The wall of the venous structure consisted of a very thin layer of collagen fibers only with a decreased quantity of fibers, and no endothelial cells, elastic laminae or muscular fibers. Fibrinous exudates had been deposited on the vascular collagen fibers, with lymphocytes and erythrocytes infiltrating the adventitia and pericardial space (Fig 5A). There were also hemosiderin-filled macrophages in the pericardium (Fig 5B). Significant atherosclerotic changes or thrombus were not observed.

The patient’s postoperative clinical course was good. Repeat CAG at 2 weeks after operation showed that the shunt flow had disappeared, but the RCA was still dilated. In addition, a small left circumflex artery–CS fistula had newly appeared, but it was not accompanied by a significant left-to-right shunt. One year after operation, under treatment with aspirin (81 mg/day) and cilostazol (200 mg/day), she remained free of symptoms. The continuous murmur was no longer audible. No ischemic change was seen on ECG (Fig 1B) and there was no evidence of PE on echocardiography.

Discussion

Although most patients with CAVF are asymptomatic, patients aged over 30 years can have symptoms. Coronary ischemia as a result of coronary steal syndrome may induce angina pectoris or, less frequently, myocardial infarction. CAVF is sometimes diagnosed as the cause of heart failure resulting from the volume overload that is secondary to the left-to-right shunt. One of the rare manifestations is endocarditis.

Rupture of the CAVF is a rare etiology of cardiac tamponade. Said et al reported that 26% of CAVF showed aneurysmal formation, but spontaneous rupture has been reported in only 3 cases6–6 Habermann et al reported an autopsy case of sudden death caused by rupture of the CAVF4 and the other 2 cases had a sudden onset of cardiac tamponade, caused by rupture of a CAVF, and the correct diagnosis was only possible after emergency surgery5.6 In those cases, severe atherosclerosis and thrombus formation were found in the resected, ruptured CAVF,4,6 Bauer et al have noted that the shear stress from the increased flow velocity and turbulence may predispose a vessel to accelerated atherosclerosis and thrombosis, resulting in occlusion of distal flow, increased intraluminal pressure, and rupture of the weakened wall.

To our knowledge, there have not been any other case reports of preoperatively diagnosed CAVF with chronic PE. The bleeding point and rupture site of the CAVF were not clear in the present case. Histopathologically, the media of the vascular wall was very thin with undeveloped elastic fibers and many erythrocytes had infiltrated the pericardium and adventitia. Furthermore, there were hemosiderin-filled macrophages in the pericardium. Based on these findings, we speculate that there had been repeated hemorrhages or the retention of blood for a long time, and we also propose the following mechanisms for the chronic PE. First, the wall of the fistula was genetically weak. Second, the shear stress from the high shunt flow dilated and weakened the vessel wall, resulting in extravasation of blood into the pericardial space and, consequently, the oozing from the aneurysmal fistula gradually increased the PE and finally caused cardiac tamponade.

Resection of the fistulous line with ligation produced an excellent clinical result, and the PE disappeared, although a newly developed fistula, originating from the left circumflex coronary artery and draining into the CS, was detected. Incomplete ablation of the CAVF can occurred in patients with multiple fistulous entry points in the receiving chamber, but in most cases these newly developed CAVFs are
hemodynamically insignificant and surgical closure is not required. The most serious complication is myocardial infarction after the repair of a CAVF with a markedly dilated coronary artery and anticoagulation treatment is necessary to prevent this complication.6,9

References